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The Problem of the Acquired Short Esophagus

Report of Eighteen Patients

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SUMMARY

A shortened esophagus is probably acquired, rather than congenital, in the great majority of cases. The process by which the shortening develops, as described by Allison and his coworkers, begins with esophageal hiatal hernia, followed by esophagitis caused by the irritation of acids from the stomach, then recurrent ulceration and healing which forms scar tissue which little by little shortens the esophagus.

Obesity and relaxation of the supporting musculotendinous structures which accompany advancing years probably are contributory factors in production of esophageal hiatal hernia. Fifteen of a series of 18 patients noted the onset of symptoms on or after the age of 45.

Roentgen examination of the esophagus and stomach is indispensable in establishing a diagnosis of acquired short esophagus. Esophagoscopic examination is even more important. In some cases endoscopic differentiation between acute inflammation and carcinoma is difficult. In such circumstances examination of a biopsy specimen taken from the gastric mucosa immediately distal to the area of inflammation or stricture may be helpful.

Results in eight patients with advanced esophageal shortening and stricture who were treated conservatively indicate that this should be tried before surgical treatment is considered. For patients with esophageal hiatal hernia accompanied by shortening of the esophagus that is just beginning to produce symptoms, early repair is indicated, since the condition is progressive and the surgical problem is much simpler in the early stages.

PEPTIC ulceration may occur at the cardioesophageal junction when there is derangement of the sphincter mechanism controlling this point of union between the esophagus and the stomach. Regurgitation of the secretions of the stomach into the lower end of the esophagus occurs most commonly as a result of esophageal hiatal hernia. In addition, it is possible in the case of esophageal hiatal hernia that the diaphragm plays a role in the concentration of gastric juice in the supradiaphragmatic portion of the stomach by offering some obstruction to complete emptying of the fundal end of the stomach, particularly when an individual so afflicted is in the horizontal position.

Esophageal hiatal hernias have been classified into three types (Figure 1):

(a) The para-esophageal hiatal hernia. In this type, the esophagus is of normal length, but a portion of the stomach has herniated into the posterior

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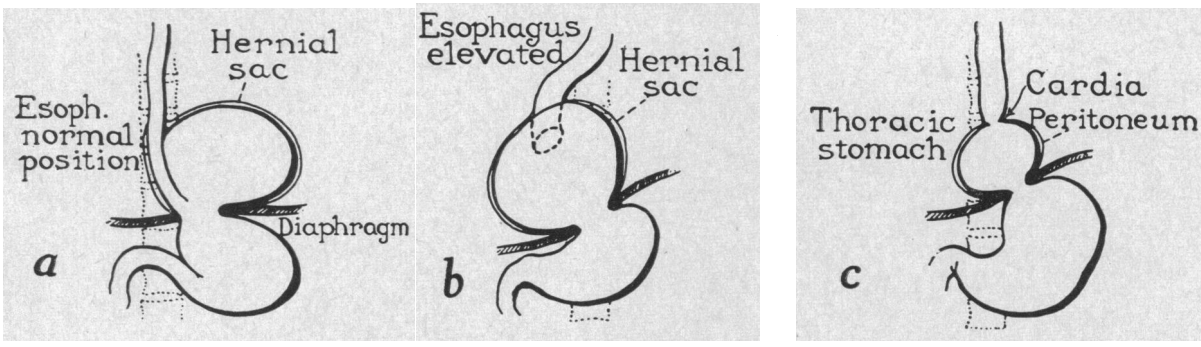

Fig. 1.—Three types of esophageal hiatal hernia; *a*, para-esophageal; *b*, sliding; *c*, short esophagus type.

From Esophageal Hiatal Hernias by A. M. Olsen, M.D., & S. W. Harrington, M.D., p.190, Journal of Thoracic Surgery, April 1948.

mediastinum beside the esophagus. This is the least common of the three types.

(b) The sliding or gastroesophageal type. This is the most common form of hiatal hernia. The esophagus is of normal length, but is elevated above the level of the diaphragm and a portion of the stomach is herniated into the posterior mediastinum. It is the belief of the author that this type of hernia may develop into the acquired short variety and therefore must be observed at regular intervals because the surgical repair of the sliding hernia is much simpler than in the case of the acquired short type.

(c) The congenital or acquired short esophagus with partial thoracic stomach.

It is probably true that the congenital short esophagus with partial thoracic stomach is an uncommon anomaly. Most persons with esophageal hiatal hernias with shortened esophagus and partial thoracic stomach acquire the shortened esophagus in the manner described by Allison and his coworkers.[1, 2] When the lower esophagus is being constantly bathed by acid from the stomach, a well-defined series of changes is found there. First there is recurrent acute esophagitis which passes on to a chronic esophagitis with recurrent acute ulceration. The acute ulcer progresses to a typical chronic ulcer. The ulcer, with its surrounding induration, produces stenosis, and above this stricture there is intense superficial inflammation. Finally, a dense fibrous stenosis is produced. Each acute exacerbation of this chronic ulcer produces additional scarring with resultant shortening of the esophagus. As the esophagus shortens, more of the stomach is pulled up into the thoracic cavity. Eventually the cardioesophageal junction may be up to the level of the inferior pulmonary vein.

Since the enlarged esophageal hiatus is the probable basis for the development of all types of esophageal hiatal hernias, some understanding of the basic embryology is in order. Olsen and Harrington[3] point out that the formation of the diaphragm coincides with the descent of the stomach from its thoracic position. The esophageal hiatus is formed by the closure of the diaphragm about the lower end of the esophagus. If the descent of the stomach is delayed, the esophageal hiatus will form about the cardiac end of the stomach. Thus the esophageal hiatus will be abnormally large. In a few instances, no further descent of the stomach will

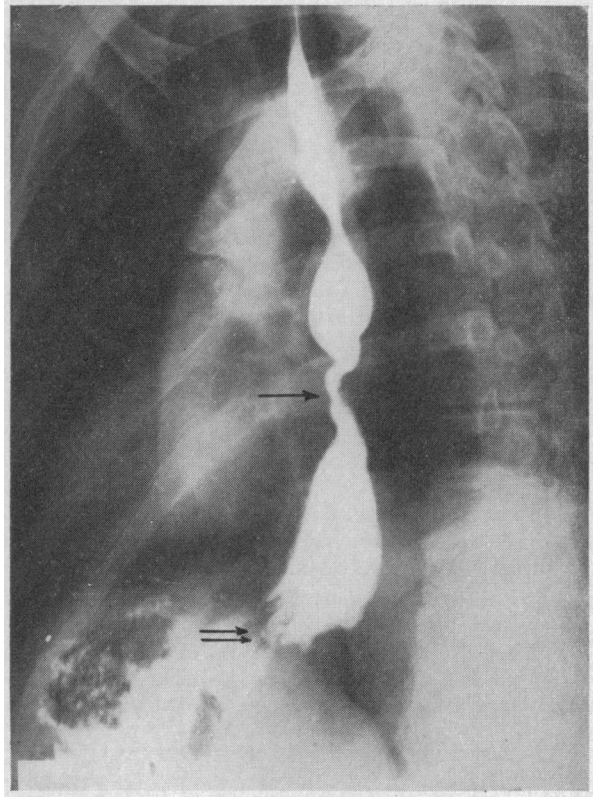

Plate I.—The single arrow shows the cardioesophageal junction at the level of the inferior pulmonary vein. The double arrow shows the hiatus opening in the diaphragm.

occur, and the result will be a true congenital short esophagus with partial thoracic stomach. However, in most instances, the stomach will continue its descent into the abdomen, thus producing an esophagus of normal length but leaving an esophageal hiatus that is large or incompetent.

Obesity and relaxation of the supporting musculotendinous structures accompany advancing years. It would seem logical that both of these conditions are important contributory factors in the production of esophageal hiatal hernia. The significance of age (Chart 1) in the etiology of the acquired short esophagus is demonstrated in the present series of 18 patients. Fifteen of these patients noted the onset of symptoms on or after the age of 45. Acrosclerosis was present in one of the three patients who had symptoms before that age, and another had had an operation to enlarge the cardioesophageal junction because of cardiospasm. A small portion of the stomach is above the diaphragm in this patient, and it is believed that an esophageal hiatal hernia was inadvertently produced when the operation to correct the cardiospasm was done.

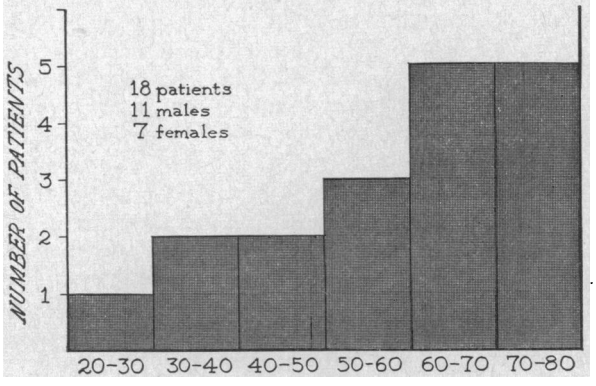

Chart 1.—Showing ages at which symptoms began in 18 patients having short esophagus with intrathoracic stomach.

There were 11 males and seven females in this group of patients who are believed to have the acquired type of esophageal hiatal hernia with shortened esophagus. This same predominance of males was present in the larger series reported by Olsen and Harrington.[3]

The majority of the 18 patients had had symptoms for several years (Chart 2). This is to be expected since dysphagia (Table 1) was an important symptom in 13 of the group. Esophagitis and acute ulceration of the esophagus express their presence by producing pain high in the epigastrium and beneath the sternum. The gastric contents may regurgitate into the throat, especially in the recumbent position, causing an extremely distressing burning sensation beneath the entire length of the sternum. Many of the patients in this group had noted the increase of the severity of their symptoms when they were in the prone, supine, or forward-bent position, and, without professional instruction, had adopted the upright position when resting or sleeping.

The chief symptoms of the chronic ulcer at the lower end of the esophagus are dysphagia and vomiting. Occasionally, stenosis is so severe that even

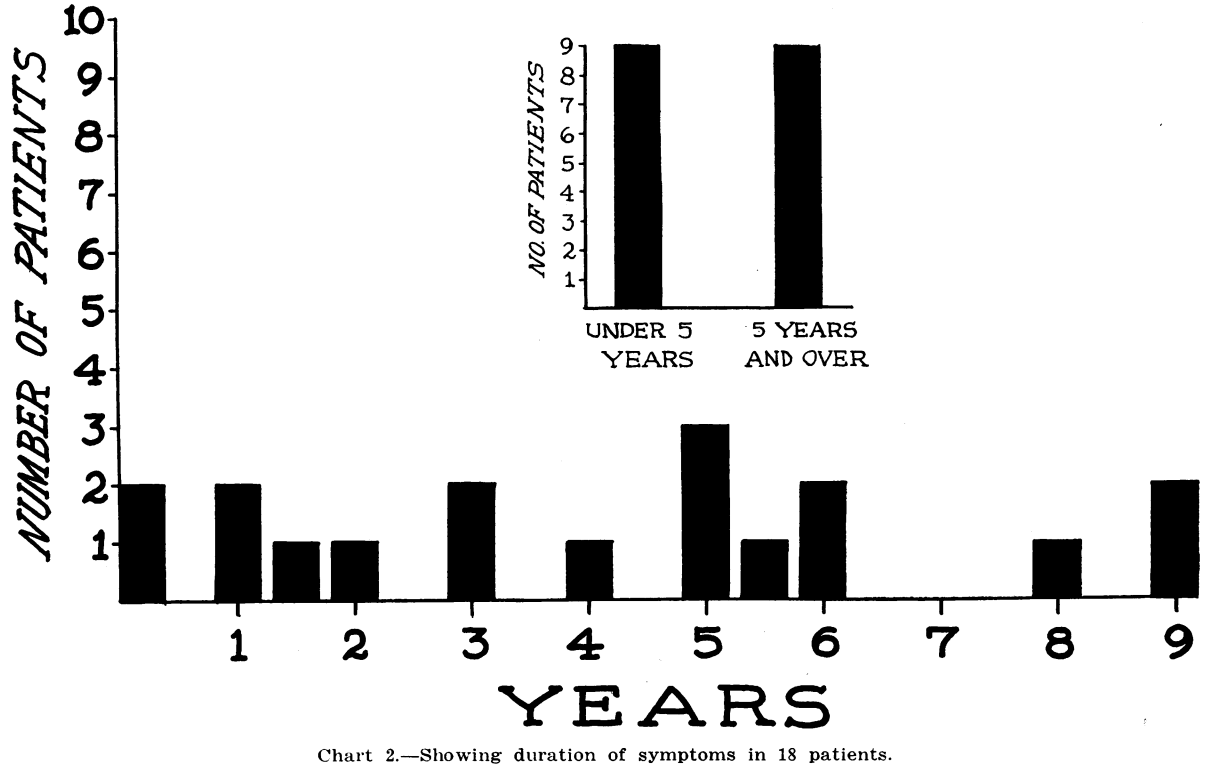

Chart 2.—Showing duration of symptoms in 18 patients.

TABLE 1

Symptoms	*No. Patients Afflicted*
Dysphagia	13
Vomiting	10
Pain	9
Melena	6
Hematemesis	6

liquids are taken with difficulty. Table 1 lists the important symptoms and their relative frequency in the group of patients observed. Bleeding into the gastrointestinal tract occurred in one-third of the group. The bleeding was severe in several of the patients, the quantity of blood passed per rectum being greatly in excess of the amount of hematemesis.

Dysphagia and vomiting are usually late symptoms but, on occasions, may appear before there is actual stricture formation. In the latter instance, these symptoms are transitory. Following the formation of a true stricture, the dysphagia and vomiting are constant and persistent until some kind of treatment is administered.

DIAGNOSIS

The diagnosis of acquired short esophagus should be established without difficulty. Roentgen examination of the esophagus and stomach is indispensable, and frequently it is necessary for the roentgenologist to examine the patient in the recumbent position to bring out the small sliding type of esophageal hiatal hernia. Not infrequently, the stricture at the cardioesophageal junction will exactly mimic the roentgen appearance of carcinoma of the esophagus. If the radiologist does not attempt to demonstrate the supradiaphragmatic stomach, then the correct diagnosis can be easily overlooked.

Esophagoscopic examination is even more important than the roentgen ray to establish the presence or absence of acquired short esophagus. Four of the 18 patients in this group were suspected of harboring other lesions until esophagoscopy demonstrated the true diagnosis. Olsen and Harrington[3] state the average length of the esophagus from the upper incisor teeth to the esophagogastric junction at the diaphragm is approximately 40.6 centimeters in the average normal adult. In the presence of acquired short esophagus, the cardioesophageal junction will be located at a higher level than in the normal. The esophagoscopic picture of the peptic ulceration of the esophagus is quite typical. There is intense injection of the esophageal mucous membrane immediately proximal to the cardioesophageal junction. Areas of leukoplakia taking on the contour outlines of shallow valleys between swollen folds of esophageal mucous membrane probably represent areas of concentration of the gastric juice as this liquid regurgitates into the lower esophagus in the absence of normal sphincter mechanism. Actual shallow or deep ulceration may be present. When the pathologic process has progressed to stricture formation, there remains evidence of acute esophagitis always located just proximal to the area of stricture.

Occasionally the peptic reaction at the lower end of the esophagus is so severe that one cannot be certain from the endoscopic picture whether the lesion is inflammatory or carcinomatous. In such an instance, a particular effort is made to obtain a biopsy specimen from the gastric mucosa situated just distal to the area of inflammation or stricture. The presence of gastric mucosa in the biopsy specimen may be the evidence that differentiates acquired short esophagus from suspected carcinoma.

TREATMENT

The treatment given to the group of patients here reported upon is summarized in Tables 2, 3, and 4. Table 2 shows that eight of the patients are either greatly improved or moderately improved under a regimen of occasional dilatations of the cardioesophageal junction by bougie or under esophagoscopic observation combined with rest, sleeping in the upright position and weight loss if indicated. The results in this group certainly commend trial of conservative management in the advanced cases of acquired short esophagus with stricture before any kind of surgical treatment is considered. The unimproved patient in this group has been an alcoholic problem for many years and probably would be resistant to any form of management.

Table 3 indicates that four patients received operative treatment. Two of this group had intractable vomiting (one of these patients has acrosclerosis and the other patient had had symptoms for nine years) due to severe stricture at the cardioesopha-

TABLE 2

Group I.—(Treatment comprised of esophagoscopy, one or several esophageal dilatations, bland diet, and sleeping in the upright position.)

Present Status	*No. of Patients*
Greatly improved	3
Greatly improved but dilatations still required	2
Moderately improved but dilatations still required	2
Moderately improved	1
No improvement	1
Did not respond to follow-up	2
Total	11

TABLE 3

Group II.

A. Repair of hiatus hernia with replacement of stomach below the diaphragm:

Greatly improved	1
Moderately improved	1

B. Resection of stenotic area of the esophagus and reanastomosis of the stomach to esophagus above the diaphragm:

Moderately improved	2
Total	4

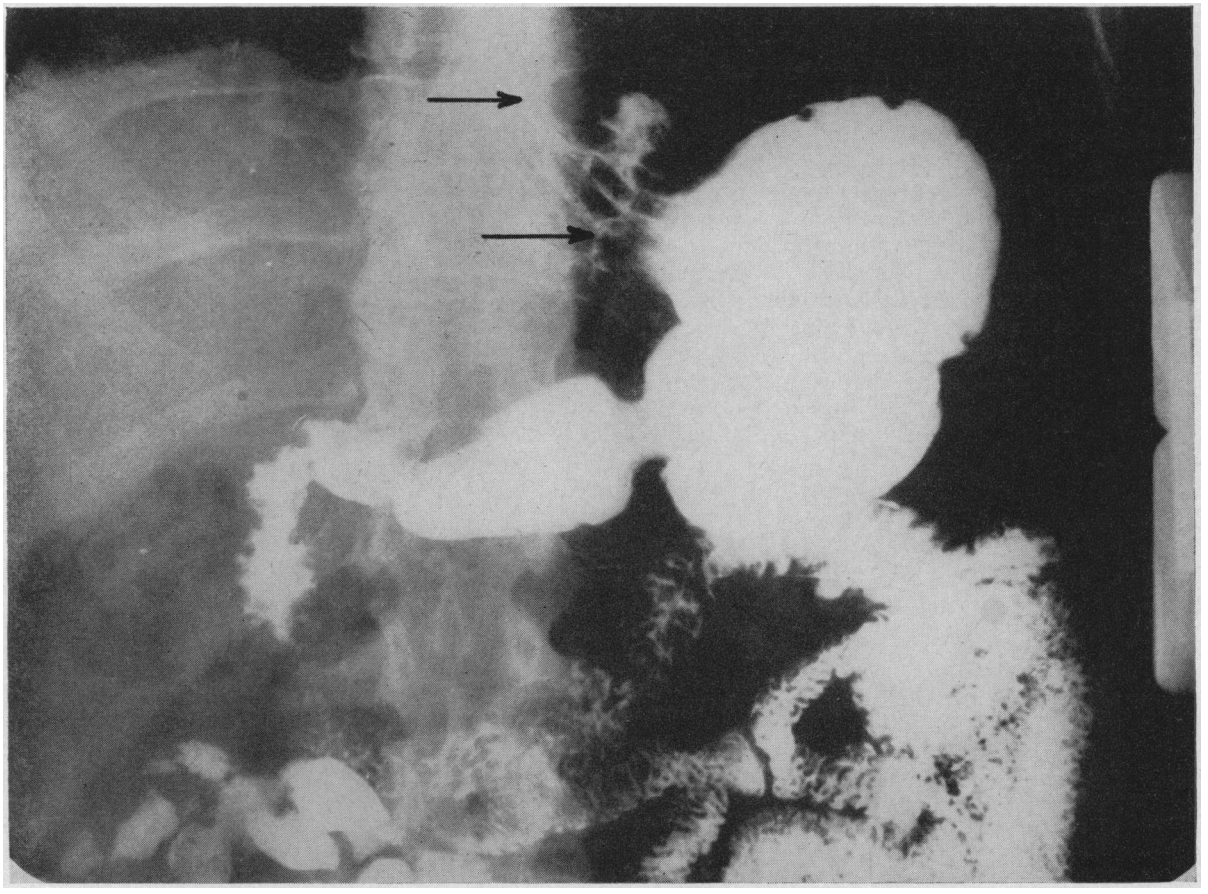

Plate II.—The two arrows demonstrate the extent of the hiatal hernia in 1941. At this stage of development, the stomach could have been replaced into the abdomen and retained there by operative repair of the hiatus. At the time of operation in 1947, the cardioesophageal junction was at the same level as seen in Plate I. It was necessary, therefore, to resect the strictured area and re-anastomose because of the distance from the diaphragm to the elevated cardioesophageal junction.

geal junction and both these patients requested operation. Resection of the strictured area of the esophagus was performed with re-anastomosis, esophagus-to-stomach, above the diaphragm.

The other two patients of this group received simple repair of the hiatal hernia through the thoracic route with replacement of the stomach below the diaphragmatic level and repair of the hiatal opening. The results in these two patients (age 49 and 61 at the time of operation) strongly suggest that surgical repair of esophageal hiatal hernia which is accompanied by early shortening of the esophagus is indicated where this condition is producing symptoms. The surgical problem is much simpler and can be more satisfactorily managed than when the cardioesophageal junction comes to occupy a position 6 to 8 centimeters above the level of the diaphragm.

Table 4 includes two patients who have died, and a third patient who is being treated outside of San Francisco. The latter patient is 53 years of age and has had symptoms for six years. A repair of the hiatal hernia was performed in 1944 with subsequent recurrence of the hernia, and the esophagus is now considerably strictured and there is a questionable ulcer in the supradiaphragmatic portion of the stomach. Experience with this patient illustrates the fact that when the cardioesophageal junction is several centimeters above the diaphragm, it is difficult to prevent recurrences in this type of hernioplasty.

When radical resection is necessary, the operations of partial esophagectomy combined with esophagoenterostomy as recommended by Allison[2] or three-quarter gastric resection as recommended recently by Wangensteen[4] are probably better surgical

TABLE 4

Group III.

A. One patient has a jejunostomy and is being prepared for esophageal resection. (This patient had a repair of her hiatus hernia in 1944 with recurrence of the hernia.)

B. One patient died in an emaciated state at the age of 82, 1½ years after first visit. Necropsy revealed ulcers at the cardioesophageal junction, lesser curvature of the stomach and duodenum. Rupture of the lesser curvature ulcer caused patient's death.

C. One patient died at the age of 82. Observed and treated for 7 years. Emaciated at time of death. In addition, had benign prostatic hypertrophy with secondary bilateral pyelonephritis.

Total 3

procedures than resection of the strictured area with re-anastomosis, esophagus-to-stomach, since in the latter operation the acid factor has not been eliminated.

The two patients who died each lived to be 82 years of age. One of these patients was observed for eight years and he would come in occasionally for an esophageal dilatation. Although this patient was emaciated at the time of death, the immediate cause of death was the presence of benign prostate hypertrophy with bilateral pyelonephritis. The other patient died in an extreme degree of emaciation. Peptic ulceration was present at the cardioesophageal junction, in the lesser curvature of the stomach and in the duodenum. The immediate cause of death in this patient was an intraperitoneal rupture of the gastric ulcer.

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